Unilateral Primary CNS Vasculitis in a Child Associated with Increased ICP and Treated with Maximal Medical Therapy and Decompressive Hemicraniectomy

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Results
- Despite maximal osmotic, sedative, and immune-directed therapies, he exhibited worsening cerebral edema and midline shift with persistently elevated intracranial pressures.

Results
- He was treated with cyclophosphamide with gradual improvement in cerebral edema.
- He underwent cranioplasty four weeks after hemicraniectomy (Hospital Day 37).

Hospital course
- Day 2 – IV methylprednisolone started
- Day 4 – Copley IPF monitor placed
- Day 5 – Pentobarbital infusion, hypertonic saline with goal Na 160s, PLEX started
- Day 10 – Had ICP spikes – went for decompressive right frontotemporoparietal hemicraniectomy with brain biopsy
- Day 12 – IV cyclophosphamide
- Day 37 – Cranioplasty
- Day 44 – Transfer to inpatient rehab

Hospital Day 16

Hospital Day 23

Hospital Day 29

Hospital Day 40 (post cranioplasty)

Conclusions / Discussion
- Our case demonstrates an unusual presentation of PACNS in a child given unilateral involvement and fulminant course.
- Hemicraniectomy should be considered in patients with medically refractory increased intracranial pressure, as it can cause irreversible morbidity and mortality.
- Management of this child involved a multidisciplinary team of providers and complex diagnostic and therapeutic decision-making, ultimately resulting in a favorable outcome.

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